Expert's comment concerning Grand Rounds case entitled “Langerhans cell histiocytosis of the atlas in an adult” (by Wo Quan Zhong, Liang Jiang, Qing Jun Ma, Zhong Jun Liu, Xiao Guang Liu, Feng Wei, Hui Shu Yuan, Geng Ting Dang)

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The authors present a case of Langerhans cell histiocytosis (LCH) of the atlas vertebra in a 26-year-old patient, and they provide us with an overview of the different presentations of the LCH. A review of the literature regarding the diagnostic workup and also of the different modalities of treatment is included. The pathologic lesion is located at the level of a lateral mass of the atlas vertebra with an obvious loss of height. The X-ray does not clearly show the importance of the lesion while it seems that the lesion is limited to the left lateral mass of atlas, and it causes a potential instability [1, 2]. We normally do not use the 3D reconstructions, yet in this specific location at the atlas level it is useful to better define the anatomic involvement. A MRI study is also useful to determine the extension of the lesion both in the soft tissue and also into the spinal canal.

Regarding the indication of a biopsy, we agree with the authors that it is a necessary step in such presentations. We would like to remind the readers that we also recommend performing a biopsy even in the typical X-ray presentations of a vertebra plana. Until now, CT and MRI are not enough to establish the diagnosis of LCH with certainty [3]. Vertebra plana may be the presentation of the different malignant etiologies including Ewing's sarcoma, osteosarcoma, and lymphoma. Some authors [4] propose a careful survey in the cases of typical vertebra plana. We prefer using needle biopsy to confirm the diagnosis. The cases where we would not consider a biopsy are those in which there are multiple bones or visceral involvement. Sometimes also in the cases of a vertebra plana that is associated with another bony involvement we may not request a biopsy. In the aforementioned circumstances, if any doubt persists regarding the diagnosis, the biopsy will be performed through the most accessible site. In all the other cases a CT-guided biopsy is our strategy.

The second point that should be discussed more in detail is that the workup of such lesions must imperatively focus together on both the bone survey and the visceral involvement. There are so many unknown issues under the title of LCH. Sometimes the lesions observed in the bone scan do not show up on the simple X-rays and in other cases it is the reverse situation that is present (visible X-rays lesions that do not light up on the bone scan). It is also a known characteristic of LCH that in the multivisceral systemic types, some pathologic sites will manifest themselves during the course of the chemotherapy. Because of the aforementioned reasons, not only an initial work up is necessary, but also a long-term surveillance is a must to detect the local recurrences.

The most controversial issue is the treatment. In the literature, the prognostic of the vertebral types is generally considered to be benign with a good end result. There have

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not been any hazardous neurological consequences after involvement of the atlas [1, 2, 4]. The spontaneous evolution will go through a healing and reconstructive process after the onset of a pathologic fracture. In children remodeling is more likely as the growth zones are little altered. For these reasons, in our department, as the basic treatment strategies, we recommend spinal immobilization with a thorough follow-up after a biopsy and a complete workup to know the extent of tumor involvement. Surgery is very rarely needed and would only be required in the cases of an extension of the lesion with the destruction of the lateral articular masses causing obvious instability [1]. If the diagnosis is certain after the biopsy the principles of surgery are simple. An instrumented stabilization after curettage and arthrodesis will achieve healing of the lesion in all cases. In the situation where would exist an intracanal extension of the lesion associated with a neurologic deficit, a treatment based upon chemotherapy and rest can be tried, but the patient must be monitored carefully. We have treated successfully a similar case with the rapid regression of the neurological signs after systemic administration of Prednisone [3]. Low dose radiation therapy seems to be overtreatment and we have abandoned this type of treatment in the isolated osseous cases. Radiotherapy has been proposed in the 1980s [5]. There is always a significant risk of malignant transformation of the lesions after radiotherapy. Greenberg has described a 4% rate of malignant transformation after irradiation for the systemic types of LCH; four patients in this series died: two of leukemia and two of thyroid cancer. This grand rounds case was a young adult patient but we would like to remind that in the case of children radiotherapy can lead to early closure of vertebral growth [6]. We recommend that for a benign lesion with a diagnosis confirmed by biopsy, knowing that the prognosis

is always favorable, radiotherapy should be as much as possible be avoided. This opinion is supported in recent published series [4, 7, 8].

In summary, a vertebral presentation of the LCH must be confirmed by a biopsy if it is a solitary lesion. A very thorough workup must look at any other bony involvement and visceral manifestations. The treatment strategy is based upon immobilization and possibly chemotherapy. Radiotherapy is in our opinion not indicated except in the very severe multivisceral cases.

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